

Annals of Neurology

Vol 52 July December 2002

Official Journal of the American Neurological Association and the Child Neurology Society



Editor

Richard T. Johnson, MD
The Johns Hopkins University
Baltimore

Founding Editor, 1977-1984

Fred Plum, MD

Editor, 1985-1992

Arthur K. Asbury, MD

Editor, 1993-1997

Robert A. Fishman, MD

Associate Editors

Ralph W. Kuncel, MD, PhD
Baltimore

David Zee, MD
Baltimore

Associate Editor for

Child Neurology

Michael V. Johnston, MD
Baltimore

Managing Editor

Sheila Rose Garrity, MS

Editorial Assistant

Andrea Greise

Publishing Staff

Shawn Morton
Vice President and Publisher, Medical
Sciences

Jessica Hickman-Beard
Senior Production Editor

Donna Lysak
Copyeditor

James G. Patis, Cunningham Associates
Advertising Representative

Annals of Neurology (ISSN 0364-5134) is published monthly by Wiley-Liss, Inc., through Wiley Subscription Services, Inc., a Wiley company for the American Neurological Association. Copyright © 2002 Wiley-Liss, Inc. All rights reserved. No part of this publication may be reproduced in any form or by any means, except as permitted under section 107 or 108 of the 1976 United States Copyright Act, without either the prior written permission of the publisher, or authorization through the Copyright Clearance Center, 222 Rosewood Drive, Danvers, MA 01923, (978) 750-8400, fax (978) 750-4470. Requests to the publisher for permission should be addressed to the Permissions Department, c/o John Wiley & Sons, Inc., 111 River Street, Hoboken, NJ 07030; (201) 748-6011, fax (201) 748-6008. E-mail: permreq@wiley.com. Periodicals postage paid at New York, NY, and at additional mailing offices.

Editorial Board

Yves Agid, MD, PhD, Paris
Rose-Mary Boustany, MD, Durham
Steven T. DeKosky, MD, Pittsburgh
Stefano Di Donato, MD, Milan
Marianne Dieterich, MD, Mainz
David J. Fink, MD, Pittsburgh
Marc Fisher, MD, Worcester
Christopher Gomez, MD, Minneapolis
Scott T. Grafton, MD, Hanover, NH
James C. Grotta, MD, Houston
Mark Hallett, MD, Bethesda
Bradley T. Hyman, MD, Boston
George Karpati, MD, Montreal
Edward M. Kaye, MD, Philadelphia
Allan I. Levey, MD, PhD, Atlanta
Roland Martin, MD, Bethesda
Solomon Moshé, MD, New York
Peter C. O'Brien, PhD, Rochester, MN
Richard K. Olney, MD, San Francisco
Louis J. Ptáček II, MD, Salt Lake City
Raymond P. Roos, MD, Chicago
Faye S. Silverstein, MD, Ann Arbor
A. Jon Stoessl, MD, Vancouver
Yoshiyuki Suzuki, MD, PhD, Otawara
Douglass M. Turnbull, MD, Newcastle
Upon Tyne

Annals of Neurology is indexed in Index Medicus, Current Contents/Clinical Medicine, Excerpta Medica, Biological Abstracts, Psychological Abstracts, and CINAHL.

Address for subscription information, orders, or changes of address: John Wiley & Sons, Inc., attn: UK Journals Admin Dept UK, 111 River Street, Hoboken, NJ 07030; (201) 748-6645 or e-mail: subinfo@wiley.com. If claims are not resolved satisfactorily, please write to Caroline Rothaug, Subscription Fulfillment and Distribution, c/o John Wiley & Sons, Inc., 111 River Street, Hoboken, NJ 07030.

Annual subscription rates: Subscription Price: (Volumes 51 and 52, 2002) Print only: \$370.00 in U.S., \$370.00 in Canada and Mexico, \$499.00 outside North America. Electronic only: \$370.00 worldwide. A combination price of \$389.00 in U.S., \$389.00 in Canada and Mexico, \$518.00 outside North America, includes the subscription in both electronic and print formats. Personal rate: \$215.00 in North America, and \$284.00 rest of world. All subscriptions containing a print element, shipped outside U.S., will be sent by air. Payment must be made in U.S. dollars drawn on a U.S. bank. Claims for undelivered copies will be accepted only after the following issue has been delivered. Prices subject to change without notice. Copies will be replaced without charge if the publisher receives a request within 90 days of the mailing date, both in the U.S. and worldwide. POSTMASTER: Send address changes to Annals of Neurology, Subscription Distribution, c/o John Wiley & Sons, Inc., 111 River Street, Hoboken, NJ 07030.

Message from the Editors

- 1 Richard T. Johnson, MD and Sheila Rose Garrity, MS

Notice

- 2 Ad Hoc Reviewers

Editorials

- 5 Hyperglycemia and Stroke Outcome: Vindication of The Ischemic Penumbra

Myron D. Ginsberg, MD

- 6 Bourneville and Taylor: A Developing Story?

Peter B. Crino, MD, PhD

Neurological Progress

- 10 Cortical Control of Spatial Memory in Humans: The Visuoculomotor Model

Charles Pierrot-Deseilligny, MD, René M. Müri, MD, Sophie Rivaud-Pechoux, PhD, Bertrand Gaymard, MD, PhD, and Christoph J. Ploner, MD

Original Articles

- 20 Acute Hyperglycemia Adversely Affects Stroke Outcome: A Magnetic Resonance Imaging and Spectroscopy Study

Mark W. Parsons, FRACP, P. Alan Barber, PhD, FRACP, Patricia M. Desmond, MD, Tracey A. Baird, MRCP, David G. Darby, PhD, FRACP, Graham Byrnes, PhD, Brian M. Tress, MD, FRACP, and Stephen M. Davis, MD, FRACP

- 29 Focal Cortical Dysplasia of Taylor's Balloon Cell Type: Mutational Analysis of the TSC1 Gene Indicates a Pathogenic Relationship to Tuberous Sclerosis

Albert J. Becker, MD, Horst Urbach, MD, Björn Scheffler, MD, Thomas Baden, Sabine Normann, Rainer Lahl, MD, Heinz W. Pannek, MD, Ingrid Tuxhorn, MD, Christian E. Elger, MD, FRCP, Johannes Schramm, MD, Otmär D. Wiestler, MD, and Ingmar Blümcke, MD

- 38 Succinate in Dystrophic White Matter: A Proton Magnetic Resonance Spectroscopy Finding Characteristic for Complex II Deficiency

Knut Brockmann, MD, Alf Bjørnstad, MD, Peter Dechent, PhD, Christoph G. Korenke, MD, Jan Smeitink, MD, J. M. Frans Trijbels, MD, Sabine Athanassopoulos, MD, Rafael Villagran, MD, Ola H. Skjeldal, MD, Ekkehard Wilichowski, MD, Jens Frahm, PhD, and Folker Hanefeld, MD

- 47 Application of the New McDonald Criteria to Patients with Clinically Isolated Syndromes Suggestive of Multiple Sclerosis

Catherine M. Dalton, MRCP, Peter A. Brex, MRCP, Katherine A. Miszkil, FRCP, Simon J. Hickman, MRCP, David G. MacManus, MSc, Gordon T. Plant, FRCP, Alan J. Thompson, FRCP, and David H. Miller, FRCP

- 54 Minocycline Markedly Protects the Neonatal Brain against Hypoxic-Ischemic Injury

Kara L. Arvin, MD, Byung H. Han, PhD, Yansheng Du, PhD, Sui-zhen Lin, MD, Steven M. Paul, MD, and David M. Holtzman, MD

- 62 Magnetization Transfer Imaging in Normal Aging, Mild Cognitive Impairment, and Alzheimer's Disease

Wiesje M. van der Flier, MA, Dominique M. J. van den Heuvel, MA, Annelies W. E. Weverling-Rijnsburger, MD, Eduard L. E. M. Bollen, MD, PhD, Rudi G. J. Westendorp, MD, PhD, Mark A. van Buchem, MD, PhD, and Huub A. M. Middelkoop, PhD

- 68 Sudomotor Testing Predicts the Presence of Neutralizing Botulinum A Toxin Antibodies

Frank Birkelein, MD, Dirk Walther, Hans Bigalke, MD, Martin Winterboller, MD, and Frank Erbguth, MD

- 74 Mechanisms of Acute Cerebral Infarctions in Patients with Middle Cerebral Artery Stenosis: A Diffusion-Weighted Imaging and Microemboli Monitoring Study

Ka Sing Wong, MD, Shan Gao, MB, Yu Leung Chan, FRCP, Tjark Hansberg, MD, Wynnne W. M. Lam, FRCP, Dirk W. Droste, MD, Richard Kay, MD, and E. Bernd Ringelstein, MD

- 82 Working Memory after Traumatic Brain Injury in Children

Harvey S. Levin, PhD, Gerri Hanten, PhD, Chung-Chi Chang, PhD, Lifang Zhang, MD, Russell Schachar, MD, Linda Ewing-Cobbs, PhD, and Jeffrey E. Max, MD

- 89 Interaction of Cognitive Aging and Memory Deficits Related to Epilepsy Surgery

Christoph Helmstaedter, PhD, Markus Reuber, MD, and Christian C. E. Elger, MD

Brief Communications

- 95 Adenine Nucleotide Translocator 1 Deficiency Associated with Sengers Syndrome

Eric Z. Jordens, PhD, Luigi Palmieri, PhD, Marjan Huizing, PhD, Lambert P. van den Heuvel, PhD, Rob C. A. Sengers, MD, PhD, Andrea Dörner, PhD, Wim Ruitenbeek, PhD, Frans J. Trijbels, PhD, Jullius Valsson, MD, Gunnlaugur Sigfusson, MD, Ferdinando Palmieri, MD, and Jan A. M. Smeitink, MD, PhD

- 99 *Angiostrongylus cantonensis* Infection Mimicking a Spinal Cord Tumor

Supinda Petjom, MD, Benjaporn Chaiwun, MD, Jongkolnee Settakorn, MD, Pannee Visrutaratna, MD, Samreung Rangdaeng, MD, and Paul S. Thorner, MD, PhD

- 102 Microscopic R2* Mapping of Reduced Brain Iron in the Belgrade Rat

Holly A. Zywickie, BS, Peter van Gelderen, PhD, James R. Connor, PhD, Joseph R. Burdo, BS, Michael D. Garrick, PhD, Kevin G. Dolan, BS, Joseph A. Frank, MD, and Jeff W. M. Bulte, PhD

- 105 Preferential Loss of Paternal 19q, but Not 1p, Alleles in Oligodendrogliomas

Marc Sanson, MD, PhD, Pascal Leuraud, MSc, Yannick Marie, MSc, Jean-Yves Delattre, MD, and Khê Hoang-Xuan, MD

- 108 Minicolumnar Pathology in Dyslexia**
Manuel F. Casanova, MD, Daniel P. Buxhoeveden, PhD,
Morris Cohen, EdD, Andrew E. Switala,
and Emil L. Roy, PhD
- 111 Anti-Hu Antibodies in Merkel Cell Carcinoma**
John E. Greenlee, MD, John D. Steffens, MD,
Susan A. Clawson, BS, Kenneth Hill, BS,
and Josep Dalmau, MD
- 115 Creutzfeldt-Jakob Disease Cluster in an Australian Rural City**
Steven Collins, MD, Alison Boyd, Post Grad Dip Gen Coun,
Ashley Fletcher, BSc, John Kaldor, PhD, Andrew Hill, PhD,
Stephen Farish, MEd, Carriona McLean, MD,
Zahid Ansari, MPH, Margaret Smith, BSc,
and Colin L. Masters, MD
- 119 In Vivo Study Indicating Loss of Intracortical Inhibition in Tumor-Associated Epilepsy**
Kerstin Irlbacher, MD, Stephan A. Brandt, MD,
and Bernd-Ulrich Meyer, MD
- 122 Germline Mosaicism of a Novel Mutation in Lysosome-Associated Membrane Protein-2 Deficiency (Danon Disease)**
Maki Takahashi, MD, Ayaka Yamamoto, MD,
Kyoko Takano, MD, Akira Sudo, MD,
Takahito Wada, MD, PhD, Yu-ichi Goto, MD, PhD,
Ichizo Nishino, MD, PhD, and Shinji Saitoh, MD, PhD

Letters

- 126 Creatine Supplementation Results in Elevated Phosphocreatine/Adenosine Triphosphate (ATP) Ratios in the Calf Muscle of Athletes but Not in Patients with Myopathies**
J. Zange, PhD, C. Kornblum, MD, K. Müller, MD,
S. Kurtscheid, MSc, H. Heck, MD, R. Schröder, MD,
T. Grehl, MD, PhD and M. Vorgerd, MD
- 126 Reply**
Mark A. Tarnopolsky, MD, PhD
- 127 Rabbit Model of Guillain-Barré Syndrome**
Irving Nachamkin, PhD
- 128 Reply**
Nobuhiro Yuki, MD, PhD, Izumi Mori, MD,
and Keiichiro Susuki, MD, PhD
- 129 Mistaken Treatment of Anterior Ischemic Optic Neuropathy with Interferon β -1a**
Jonathan C. Horton, MD, PhD

Regular Departments

130 Books

NO 2 AUGUST 2002

Editorial

- 133 New Neurons for Aging Brains**
Guy M. McKhann, MD

Original Articles

- 135 Neuroplasticity in Old Age: Sustained Fivefold Induction of Hippocampal Neurogenesis by Long-term Environmental Enrichment**
Gerd Kempermann, MD, Daniela Gast,
and Fred H. Gage, PhD
- 144 Pallidal Stimulation for Parkinsonism: Improved Brain Activation during Sequence Learning**
Masafumi Fukuda, MD, Maria Felice Ghilardi, MD,
Maren Carbon, MD, Vijay Dhawan, PhD, Yilong Ma, PhD,
Andrew Feigin, MD, Marc J. Mentis, MD,
Claude Ghez, MD, and David Eidelberg, MD
- 153 A Nonischemic Forearm Exercise Test for McArdle Disease**
Pedram Kazemi-Esfarjani, MD, Elwira Skomrowska, MD,
Tina Dysgaard Jensen, BS, Ronald G. Haller, MD,
and John Vissing, MD, PhD
- 160 Gene Transfer of HSP72 Protects Cornu Ammonis 1 Region of the Hippocampus Neurons from Global Ischemia: Influence of Bcl-2**
Stephen Kelly, PhD, Zhijian J. Zhang, MD, PhD,
Heng Zhao, PhD, Lijun Xu, MD,
Rona G. Giffard, MD, PhD, Robert M. Sapolsky, PhD,
Midori A. Yenari, MD, and Gary K. Steinberg, MD, PhD

- 168 Early Inflammation and Dementia: A 25-Year Follow-up of the Honolulu-Asia Aging Study**
Reinhold Schmidt, MD, Helena Schmidt, MD,
J. David Curb, MD, Kamal Masaki, MD,
Lon R. White, MD, and Lenore J. Launer, PhD
- 175 Peripheral F₂-Isoprostanes and F₄-Neuroprostanes Are Not Increased in Alzheimer's Disease**
Thomas J. Montine, MD, PhD, Joseph F. Quinn, MD,
Dejan Milatovic, PhD, Lisa C. Silbert, MD,
Theresa Dang, BS, Stephanie Sanchez, BS, Erin Terry, MS,
L. Jackson Roberts II, MD, Jeffrey A. Kaye, MD,
and Jason D. Morrow, MD
- 180 Differences in Membrane Properties of Axonal and Demyelinating Guillain-Barré Syndromes**
Satoshi Kuwabara, MD, Kazue Ogawara, MD,
Jia-Ying Sung, MD, Masahiro Mori, MD,
Kazuaki Kanai, MD, Takamichi Hattori, MD,
Nobuhiro Yuki, MD, Cindy S.-Y. Lin, BE, MEngSc,
David Burke, MD, DSc, and Hugh Bostock, PhD, FRS
- 188 Premotor Cortex Is Involved in Restoration of Gait in Stroke**
Ichiro Miyai, MD, PhD, Hajime Yagura, MD,
Ichiro Oda, PhD, Ikuo Konishi, PhD, Hideo Eda, PhD,
Tsunehiro Suzuki, MD, PhD, and Kisou Kubota, MD, PhD
- 195 Cofactors of Mitochondrial Enzymes Attenuate Copper-Induced Death In Vitro and In Vivo**
Christian T. Sheline, PhD, Eric H. Choi,
Jeong-Sook Kim-Han, PhD, Laura L. Dugan, MD,
and Dennis W. Choi, MD, PhD

205 Novel Antibodies to Synuclein Show Abundant Striatal Pathology in Lewy Body Diseases

John E. Duda, MD, Benoit I. Giasson, PhD,
Meghann E. Mabon, BA,
Virginia M.-Y. Lee, PhD,
and John Q. Trojanowski, MD, PhD

211 Mutations of Mitochondrial DNA Polymerase γ A Are a Frequent Cause of Autosomal Dominant or Recessive Progressive External Ophthalmoplegia

Eleonora Lamantea, MSc,
Valeria Tiranti, PhD,
Andreina Bordini, BSc,
Antonio Toscano, MD,
Francesco Bono, MD, Serena Servidei, MD,
Alex Papadimitriou, MD, Hans Spelbrink, PhD,
Laura Silvestri, PhD,
Giorgio Casari, PhD,
Giacomo P. Comi, MD,
and Massimo Zeviani, MD, PhD

Brief Communications

220 Progressive Loss of Cardiac Sympathetic Innervation in Parkinson's Disease

Sheng-Ting Li, MD, PhD,
Raghuvier Dendi, MD,
Courtney Holmes, CMT,
and David S. Goldstein, MD, PhD

224 No Mutation in the *TRKA* (*NTRK1*) Gene Encoding a Receptor Tyrosine Kinase for Nerve Growth Factor in a Patient with Hereditary Sensory and Autonomic Neuropathy Type V

Ennio Toscano, MD, PhD,
Alessandro Simonati, MD,
Yasuhiro Indo, MD, PhD,
and Generoso Andria, MD

227 X-Linked Creatine Deficiency Syndrome: A Novel Mutation in Creatine Transporter Gene *SLC6A8*

Alberto Bizzi, MD, Marianna Bugiani, MD,
Gajja S. Salomons, PhD,
Donald H. Hunneman, PhD,
Isabella Moroni, MD, Margherita Estienne, MD,
Ugo Danesi, PhD, Cornelis Jakobs, PhD,
and Graziella Uziel, MD

231 Vertebrobasilar Stroke as a Late Complication of a Blalock-Taussig Shunt

Philippe Gailloud, MD, Argye Hillis, MD,
Bruce Perler, MD, and Kieran J. Murphy, MD

234 Cerebral X-Linked Adrenoleukodystrophy in a Girl with Xq27-Ter Deletion

Eli Hershkowitz, MD, Ginat Narkis, BA,
Zamir Shorer, MD, Ann B. Moser, BA,
Paul A. Watkins, MD, PhD,
Hugo W. Moser, MD,
and Esther Manor, PhD

237 A Novel Mutation in the Deoxyguanosine Kinase Gene Causing Depletion of Mitochondrial DNA

Jan-Willem Taanman, PhD, Ihab Kateeb, MD,
Ania C. Muntau, MD, Michaela Jaksch, MD,
Nadine Cohen, MD,
and Hanna Mandel, MD

240 Dyskinesias and Grip Control in Parkinson's Disease Are Normalized by Chronic Stimulation of the Subthalamic Nucleus

Roland Wenzelburger, MD, Bao-Rong Zhang, MD,
Meike Poepping, MD,
Bettina Schrader, MD,
Dieter Müller, PhD,
Florian Kopper, MD,
Urban Fietzek, MD,
Hubertus M. Mehdorn, PhD,
Günther Deuschl, PhD,
and Paul Krack, PhD

243 Demonstration of Acute Ischemic Lesions in the Fetal Brain by Diffusion Magnetic Resonance Imaging

Cristina Baldoli, MD, Andrea Righini, MD,
Cecilia Parazzini, MD, Giuseppe Scotti, MD,
and Fabio Triulzi, MD

247 Selective Loss of Cholinergic Sudomotor Fibers Causes Anhidrosis in Ross Syndrome

Claudia Sommer, MD, Thies Lindenlaub, MD,
Detlef Zillikens, MD, Klaus V. Toyka, MD,
and Markus Naumann, MD

251 Normokalemic Periodic Paralysis Revisited: Does It Exist?

Patrick F. Chinnery, PhD, MRCP,
Timothy J. Walls, MD, FRCP,
Michael G. Hanna, MD, MRCP, David Bates, MA, FRCP,
and Peter R. W. Fawcett, BSc, FRCP

253 Human Antibodies against Amyloid β Peptide: A Potential Treatment for Alzheimer's Disease

Richard Dodel, MD, Harald Hampel, MD,
Candan Depboylu, MD, Suizhen Lin, MD, Feng Gao, MD,
Sabine Schock, MD, Steffi Jäckel, MD,
Xing Wei, MD, Katharina Buerger, MD,
Christine Höft, BSc,
Bernhard Hemmer, MD,
Hans-Jürgen Möller, MD, Martin Farlow, MD,
Wolfgang H. Oertel, MD,
Norbert Sommer, MD,
and Yansheng Du, PhD

Letters

257 Role of SCA2 Mutations in Early- and Late-Onset Dopa-Responsive Parkinsonism

Norman Kock, MD, Birgitt Müller, MD,
Peter Vieregge, MD, Peter P. Pramstaller, MD,
Karen Marder, MD, MPH,
Giovanni Abbruzzese, MD, Paolo Martinelli, MD,
Anthony E. Lang, MD, Helfried Jacobs, MD,
Johann Hagenah, MD, Juliette Harris, MS, PhD,
Helen Meija-Santana, MS, Stanley Fahn, MD,
Katja Hedrich, MS, Martin Kann, BS,
Ulrike Gehlken, BS, Biljana Culjkovic, PhD,
Eberhard Schwinger, MD, Zbigniew K. Wszolek, MD,
Christine Zühlke, PhD,
and Christine Klein, MD

258 Reply

Din-E Shan, MD, PhD and Bing-Wen Soong, MD, PhD

Regular Departments

259 Books

Editorials

- 261 **The Ever-Stimulating Association of Smoking and Coffee and Parkinson's Disease**
Karen Marder, MD, MPH
and Giancarlo Logroscino, MD, PhD
- 263 **Tauists, Baptists, Syners, Apostates, and New Data**
John Q. Trojanowski, MD, PhD

Neurological Progress

- 266 **Cell Transplantation for Stroke**
Sean I. Savitz, MD, Daniel M. Rosenbaum, MD,
Jonathan H. Dinsmore, PhD, Lawrence R. Wechsler, MD,
and Louis R. Caplan, MD

Original Articles

- 276 **A Meta-analysis of Coffee Drinking, Cigarette Smoking, and the Risk of Parkinson's Disease**
Miguel A. Hernán, MD, DrPH,
Bahi Takkouche, MD, PhD,
Francisco Caamaño-Isorna, PharmD,
and Juan J. Gestal-Otero, MD, PhD
- 285 **Astrocyte-Specific *TSCI* Conditional Knockout Mice Exhibit Abnormal Neuronal Organization and Seizures**
Erik J. Uhlmann, MD, Michael Wong, MD, PhD,
Rebecca L. Baldwin, BS, M. Livia Bajenaru, PhD,
Hiroaki Onda, PhD, David J. Kwiatkowski, MD, PhD,
Kelvin Yamada, MD, and David H. Gutmann, MD, PhD
- 297 **Complex Segregation Analysis of Restless Legs Syndrome Provides Evidence for an Autosomal Dominant Mode of Inheritance in Early Age at Onset Families**
Juliane Winkelmann, MD, Bertram Muller-Myhsok, MD,
Hans-Ulrich Wittchen, PhD, Bettina Hock, Muriel Prager,
Hildegard Pfister, Andreas Stroble, MD,
Ilonka Eisenbr, MD, Martin Dichgans, MD,
Thomas Gasser, MD, and Claudia Trenkwalder, MD
- 303 **Status Epilepticus in a Population-Based Cohort with Childhood-Onset Epilepsy in Finland**
Matti Sillanpää, MD, PhD, and Shlomo Shinnar, MD, PhD
- 311 **Mitochondrial DNA Depletion and *dGK* Gene Mutations**
Leonardo Salviati, MD, Sabrina Sacconi, MD,
Michelangelo Mancuso, MD, David Otaegui, BS,
Pilar Camaró, BS, Alberto Marina, PhD,
Simon Rabinowitz, MD, PhD, Rebecca Shiffman, MD,
Karen Thompson, MD, Claire M. Wilson, MD,
Annette Feigenbaum, MD, Ali B. Naini, PhD,
Michio Hirano, MD, Eduardo Bonilla, MD,
Salvatore DiMauro, MD, and Tuan H. Vu, MD
- 318 **Hyperthermia Induces Age-Dependent Changes in Rat Hippocampal Excitability**
Michelle T. Liebrechts, MSc,
Richard S. McLachlan, MD, FRCPC,
and L. Stan Leung, PhD

- 327 **Exploring White Matter Tracts in Band Heterotopia Using Diffusion Tractography**
Sofia H. Eriksson, MD, Mark R. Symms, PhD,
Fergus J. Rugg-Gunn, MD, Philip A. Boulby, PhD,
Claudia A. M. Wheeler-Kingshott, PhD,
Garth J. Barker, PhD, John S. Duncan, DM,
and Geoffrey J. M. Parker, PhD
- 335 **Periventricular Cerebral White Matter Lesions Predict Rate of Cognitive Decline**
Jan Cees de Groot, MD, Frank-Erik de Leeuw, MD,
Matthijs Oudkerk, MD, Jan van Gijn, FRCPE,
Albert Hofman, MD, Jellemer Jolles, PhD,
and Monique M. B. Breteler, MD
- 342 **Midodrine in Neurally Mediated Syncope: A Double-Blind, Randomized, Crossover Study**
Horacio Kaufmann, MD, Daniela Saadia, MD,
and Andrei Voustianiouk, PhD
- 346 **Treatment with Simvastatin in Normocholesterolemic Patients with Alzheimer's Disease: A 26-Week Randomized, Placebo-Controlled, Double-blind Trial**
Mikael Simons, MD, Frank Schwärzler, MD,
Dieter Lütjohann, PhD, Klaus von Bergmann, MD,
Konrad Beyreuther, PhD, Johannes Dichgans, MD,
Henning Wormstall, MD, Tobias Hartmann, PhD,
and Jörg B. Schulz, MD

Brief Communications

- 351 **Niemann-Pick Type C Disease: Accelerated Neurofibrillary Tangle Formation and Amyloid β Deposition Associated with Apolipoprotein E $\epsilon 4$ Homozygosity**
Yuko Saito, MD, PhD, Kinuko Suzuki, MD,
Eiji Nanba, MD, PhD, Toshiyuki Yamamoto, MD, PhD,
Kousaku Ohno, MD, PhD,
and Shigeo Murayama, MD, PhD
- 355 **Spontaneous Mutations in the Prion Protein Gene Causing Transmissible Spongiform Encephalopathy**
Ayush Dagvadorj, MD, Robert B. Petersen, PhD,
Hee Suk Lee, MD, Larisa Cervenakova, MD,
Alexey Shatunov, PhD, Herbert Budka, MD,
Paul Brown, MD, Pierluigi Gambetti, MD,
and Lev G. Goldfarb, MD
- 359 **Exclusion Mapping of the Genetic Predisposition for Cervical Artery Dissections by Linkage Analysis**
Caspar Grond-Ginsbach, PhD, Bernhard Klima, MD,
Ralf Weber, MD, Johannes Striegel, MD,
Christine Fischer, MS, Werner Hacke, MD,
Tobias Brandt, MD, and Ingrid Hauser, MD
- 364 **LGII Is Mutated in Familial Temporal Lobe Epilepsy Characterized by Aphasic Seizures**
Wenli Gu, PhD, Eyler Brodtkorb, MD, PhD,
and Ortrud K. Steinlein, MD
- 367 **Impaired Chemosensitivity to Hypoxia Is a Marker of Multiple System Atrophy**
Takehide Tsuda, MD, PhD, Hiroshi Onodera, MD, PhD,
Shinichi Okabe, MD, PhD, Yoshihiro Kikuchi, MD, PhD,
and Yasuto Itoyama, MD, PhD

- 371 **Increased Urine Phenylethylamine after Methylphenidate Treatment in Children with ADHD**
Akira Kusaga, MD, Yushiro Yamashita, MD, Tetsuya Koeda, MD, Michio Hiratani, MD, Mika Kaneko, MD, Shigeto Yamada, MD, and Toyojiro Matsuishi, MD

- 374 **Leigh-like Encephalopathy Complicating Leber's Hereditary Optic Neuropathy**
Benoît Funalot, MD, Pascal Reynier, MD, PhD, Alain Vighetto, MD, Danièle Ranoux, MD, Jean-Paul Bonnefont, MD, PhD, Catherine Godinot, PhD, Yves Malthiery, MD, PhD, and Jean-Louis Mas, MD

- 378 **Clasmatodendrosis Correlating with Periventricular Hyperintensity in Mixed Dementia**
Demetrios J. Sahlas, MD, MSc, Juan M. Bilbao, MD, Richard H. Swartz, PhD, and Sandra E. Black, MD

Letters

- 382 **No Evidence for Individual Blood-Brain Barrier Phenylalanine Transport to Influence Clinical Outcome in Typical Phenylketonuria Patients**
Joachim Pietz, MD, André Rupp, PhD, Peter Burgard, PhD, Chris Boesch, MD, PhD, Roland Kreis, PhD

- 383 **Reply**
Josef Weglage, MD, PhD, Dirk Wiedermann, PhD, Reinhold Feldmann, PhD, Kurt Ullrich, MD, and Harald E. Möller, PhD

- 384 **Mild Cognitive Impairment and the Cholinergic Hypothesis: A Very Different Take on Recent Data**
Martin Sarter, PhD, and John P. Bruno, PhD

Regular Departments

- 386 **Books**

NO 3 SEPTEMBER 2002 SUPPLEMENT 1

Neurology Outcomes Research: Current Science and Future Directions

- S4 Program
S13 Abstracts

127th Annual Meeting of the American Neurological Association

- S1 Program
S19 Abstracts

31st Annual Meeting of the Child Neurology Society

- S98 Program
S112 Abstracts

NO 4 OCTOBER 2002

Editorial

- 389 **Gliomatosis Cerebri: Molecular Pathology and Clinical Course**
Peter C. Burger, MD

Original Articles

- 390 **Gliomatosis Cerebri: Molecular Pathology and Clinical Course**
Ulrich Herrlinger, MD, Jörg Felsberg, MD, Wilhelm Küker, MD, Antje Bornemann, MD, Ludwig Plaschwil, MD, Christiane B. Knobbe, MD, Herwig Strik, MD, Wolfgang Wick, MD, Richard Meyermann, MD, Johannes Dichgans, MD, Michael Bamberg, MD, Guido Reifenberger, MDAU, and Michael Weller, MD

- 400 **Assessment of Different Treatment Failure Criteria in a Cohort of Relapsing-Remitting Multiple Sclerosis Patients Treated with Interferon β : Implications for Clinical Trials**

Jordi Río, MD, Carlos Nos, MD, Mar Tintoré, MD, Cecilia Borrás, MD, Ingrid Galán, MD, Manuel Comabella, MD, and Xavier Montalban, MD

- 407 **Interictal High-Frequency Oscillations (80–500Hz) in the Human Epileptic Brain: Entorhinal Cortex**

Anatol Bragin, PhD, Charles L. Wilson, PhD, Richard J. Staba, PhD, Mark Reddick, MS, Itzhak Fried, MD, PhD, and Jerome Engel, Jr., MD, PhD

416 Valproic Acid Treatment Results in Increased Accumulation of Prion Proteins

Gideon M. Shaked, PhD,
Roni Engelstein, BSc,
Inbal Avraham, BSc,
Hannah Rosenmann, PhD,
and Ruth Gabizon, PhD

421 Treatment of Multiple Sclerosis with the Pregnancy Hormone Estriol

Nancy L. Sicotte, MD, Stephanie M. Liva, PhD,
Rochelle Klutch, RN, Paul Pfeiffer, BS,
Seth Bouvier, BS, Sylvia Odesa, BS,
T. C. Jackson Wu, MD, PhD,
and Rhonda R. Voskuhl, MD

429 Transient Central Nervous System White Matter Abnormality in X-Linked Charcot-Marie-Tooth disease

Henry L. Paulson, MD, PhD, James Y. Garbern, MD, PhD,
Timothy F. Hoban, MD, Karen M. Krajewski, MS,
Richard A. Lewis, MD,
Kenneth H. Fischbeck, MD,
Robert I. Grossman, MD,
Robert Lenkinski, PhD,
John A. Kamholz, MD, PhD,
and Michael E. Shy, MD

435 Instability of A Premutation Allele in Homozygous Patients With Myotonic Dystrophy Type 1

Claudia Abbruzzese, PhD, Sandro Costanzi Porrini, PhD,
Bruno Mariani, PhD, Fiona K. Gould, MSc,
John P. Mcabney, Darren G. Monckton, PhD,
Tetsuo Ashizawa, MD, and Manlio Giacomelli, MD

442 *Wld⁺* Mice Are Resistant to Paclitaxel (Taxol) Neuropathy

Min Sheng Wang, PhD, Albert A. Davis, BS,
Deborah G. Culver, MS, and Jonathan D. Glass, MD

448 Evidence That Accumulation of Ceramides and Cholesterol Esters Mediates Oxidative Stress-Induced Death of Motor Neurons in Amyotrophic Lateral Sclerosis

Roy G. Cutler, MS, Ward A. Pedersen, PhD,
Simonetta Camandola, PhD, Jeffrey D. Rothstein, MD,
and Mark P. Mattson, PhD

458 Imaging the Metabolic Footprint of Glut1 Deficiency on the Brain

Juan M. Pascual, MD, PhD, Ronald L. van Heertum, MD,
Dong Wang, MD, Kristin Engelstad, BS,
and Darryl C. De Vivo, MD

465 Aggregation of Actin and Cofilin in Identical Twins with Juvenile-Onset Dystonia

Marla Gearing, PhD, Jorge L. Juncos, MD,
Vincent Procaccio, MD, PhD, Claire-Anne Gutekunst, PhD,
Elaine M. Marino-Rodriguez, MS, Kimberly A. Gyure, MD,
Shoichiro Ono, PhD, Robert Santoian, HTL(ASCP),
Nicolas S. Krawiecki, MD,
Douglas C. Wallace, PhD,
and Bruce H. Wainer, MD, PhD

477 A PLP Splicing Abnormality Is Associated with an Unusual Presentation of PMD

Grace M. Hobson, PhD, Zhong Huang, MD,
Karen Sperle, MS,
Deborah L. Stabley, BS,
Harold G. Marks, MD,
and Franca Cambi, MD

Brief Communications

489 Myoclonus-Dystonia Syndrome: ϵ -Sarcoglycan Mutations and Phenotype

Friedrich Asmus, MD, Alexander Zimprich, MD,
Sophie Tezenas du Montcel, MD, Christian Kabus, MD,
Günther Deuschl, MD, Andreas Kupsch, MD,
Ulf Ziemann, MD, Mirna Castro,
Andrea A. Kühn, MD, Tim M. Strom, MD,
Marie Vidailhet, MD,
Kailash P. Bhatia, MD,
Alexandra Dürr, MD, PhD,
Nicholas W. Wood, PhD, FRCP,
Alexis Brice, MD, and Thomas Gasser, MD

492 Preserved Visual Function in a Case of Occipitoparietal Microgyria

Pascal Zesiger, PhD, Daniel Kiper, PhD,
Philippe Maeder, MD, Thierry Deonna, MD,
and Giorgio M. Innocenti, MD

498 Involvement of Lysosomes in the Pathogenesis of CAG Repeat Diseases

Mitsunori Yamada, MD, PhD, Shoji Tsuji, MD, PhD,
and Hitoshi Takahashi, MD, PhD

503 Quinacrine Does Not Prolong Survival in a Murine Creutzfeldt-Jakob Disease Model

Steven J. Collins, MD, Victoria Lewis, BSc (Hons),
Marcus Brazier, BSc (Hons),
Andrew F. Hill, PhD,
Ashley Fletcher, BSc, and Colin L. Masters, MD

506 Cree Leukoencephalopathy and CACH/VWM Disease Are Allelic at the *EIF2B5* Locus

Anne Fogli, PhD, Kondi Wong, MD,
Eleonore Eymard-Pierre, PhD, Jack Wenger, BSc,
John-Paul Bouffard, MD, Ehud Goldin, PhD,
Deborah N. Black, MD,
Odile Boespflug-Tanguy, MD, PhD,
and Raphael Schiffmann, MD

511 An $R^{5L}T$ Mutation in a Subject with a Progressive Supranuclear Palsy Phenotype

Parvoneh Poorkaj, PhD, Nancy A. Muma, PhD,
Victoria Zhukareva, PhD, Elizabeth J. Cochran, MD,
Kathleen M. Shannon, MD, Howard Hurtig, MD,
William C. Koller, MD,
Thomas D. Bird, MD,
John Q. Trojanowski, MD, PhD,
Virginia M.-Y. Lee, PhD,
and Gerard D. Schellenberg, PhD

- 516 A Novel Mutation in the GNE Gene and a Linkage Disequilibrium in Japanese Pedigrees**
Aki Arai, MD, Keiko Tanaka, MD, Takeshi Ikeuchi, MD, Shuichi Igarashi, MD, Hisashi Kobayashi, MD, Tomoya Asaka, MD, Hidetoshi Date, MS, Masaaki Saito, MD, Hajime Tanaka, MD, Sari Kawasaki, MD, Eiichi Uyama, MD, Hidehiro Mizusawa, MD, Nobuyoshi Fukuhara, MD, and Shoji Tsuji, MD

History of Neurology

- 520 Alzheimer's Second Patient: Johann F. and His Family**
Hans H. Klünemann, MD, Wolfgang Fronhöfer, Herbert Wurster, PhD, Wolfgang Fischer, MD, Bernd Ibach, MD, and Helmfried E. Klein, MD

Letters

- 524 Linkage of Parkinsonism and Alzheimer's Disease with Lewy Body Pathology to Chromosome 12**
William K. Scott, PhD, Jeffery M. Vance, PhD, MD, Jonathan L. Haines, PhD, and Margaret A. Pericak-Vance, PhD

- 524 Reply**
Manabu Funayama, MS, Kazuko Hasegawa, MD, PhD, and Fumiya Obata, PhD

- 524 Interleukin-12 p40 Polymorphism and Susceptibility to Multiple Sclerosis**
Iraide Alloza, MSc, Shirley Heggarty, PhD, An Goris, MSc, Colin A. Graham, PhD, Bénédicte Dubois, MD, PhD, Gavin McDonnell, MD, Stanley A. Hawkins, FRCP, Herwig Carton, MD, PhD, and Koen Vandenbroeck, PhD

- 525 Concerns with the Use of T2* Signal Intensity to Differentiate Hippocampal Subregions**
David G. Darby, FRACP, and Brain M. Tress, FRACP

- 526 Reply**
Scott A. Small, MD

Regular Departments

- 527 Books**

NO 5 NOVEMBER 2002

Editorials

- 529 Leber Hereditary Optic Neuropathy: A Nuclear Solution of a Mitochondrial Problem**
Nils-Göran Larsson, MD, PhD
- 530 What Does It Mean to Be a Carrier for a Metabolic Disease?**
Edward M. Kaye, MD
- 531 Polar Bear Liver, Vitamin A, Aquaporins, and Pseudotumor Cerebri**
Robert A. Fishman, MD

- 549 A Susceptibility Gene for Late-Onset Idiopathic Parkinson's Disease**
Andrew A. Hicks, PhD, Hjörvar Pétursson, BS, Thorlákur Jónsson, PhD, Hreinn Stefánsson, PhD, Hrefna S. Jóhannsdóttir, BSc, Jesus Sainz, PhD, Michael L. Frigge, PhD, Augustine Kong, PhD, Jeffrey R. Gulcher, MD, PhD, Kári Stefánsson, MD, PhD, and Sigurlaug Sveinbjörnsdóttir, MD

- 556 Long-term Amygdalohippocampal Stimulation for Refractory Temporal Lobe Epilepsy**
Kristl Vonck, MD, Paul Boon, MD, PhD, Erik Achten, MD, PhD, Jacques De Reuck, MD, PhD, and Jacques Caemaert, MD, PhD

- 566 Eubarc Hyperoxemia and Experimental Cerebral Infarction**
Erin P. Flynn, MSc, and Roland N. Auer, MD, PhD

- 573 The Structural Consequences of Newly Diagnosed Seizures**
Rebecca S. N. Liu, BSc, MRCP, Louis Lemieux, PhD, Gail S. Bell, MRCP, Sanjay M. Sisodiya, PhD, MRCP, Philippa A. Bartlett, DCR, Simon D. Shorvon, MD, FRCP, Josemir W. A. Sander, PhD, MRCP, and John S. Duncan, DM, FRCP

- 581 Disruption of the Proprioceptive Mapping in the Medial Wall of Parkinsonian Monkeys**
Ludovic Escola, PhD, Thomas Michelet, MS, Gaëlle Douillard, MS, Dominique Guehl, MD, PhD, Bernard Bioulac, MD, PhD, and Pierre Burbaud, MD/PhD

Original Articles

- 534 Rescue of a Mitochondrial Deficiency Causing Leber Hereditary Optic Neuropathy**
John Guy, MD, Xiaoping Qi, MD, Francesco Pallotti, MD, PhD, Eric A. Schon, PhD, Giovanni Manfredi, MD, Valerio Carelli, MD, PhD, Andrea Martinuzzi, MD, PhD, William W. Hauswirth, PhD, and Alfred S. Lewin, PhD
- 543 Quantitative CSF PCR in Epstein-Barr Virus Infections of the Central Nervous System**
Adriana Weinberg, MD, Shaobing Li, MD, Megan Palmer, MD, and Kenneth L. Tyler, MD

588 Abnormal Asymmetry in Language Association Cortex in Autism

Martha R. Herbert, MD, PhD, Gordon J. Harris, PhD, Kristen T. Adrien, BA, David A. Ziegler, BS, Nikos Makris, MD, PhD, Dave N. Kennedy, PhD, Nicholas T. Lange, PhD, Chris F. Chabris, PhD, Anna Bakardjiev, MD, James Hodgson, PhD, Masanori Takeoka, MD, Helen Tager-Flusberg, PhD, and Verne S. Caviness, Jr., MD

597 p53 Inhibitors Preserve Dopamine Neurons and Motor Function in Experimental Parkinsonism

Wenzhen Duan, PhD, Xiaoxiang Zhu, PhD, Bruce Ladenheim, PhD, Qian-sheng Yu, PhD, Zhihong Guo, PhD, Jon Oyler, MS, Roy G. Cutler, MS, Jean Lud Cadet, MD, Nigel H. Greig, PhD, and Mark P. Mattson, PhD

607 Motor Cortex Activation Is Preserved in Patients with Chronic Hemiplegic Stroke

Steven C. Cramer, MD, Angela Mark, MM, Kristin Barquist, Hoang Nhan, Keith C. Stegbauer, BS, Robert Price, MSME, Kathleen Bell, MD, Ib R. Odderson, MD, PhD, Peter Esselman, MD, and Kenneth R. Maravilla, MD

617 Intravenous TAT-Bcl-X_L Is Protective after Middle Cerebral Artery Occlusion in Mice

Ertugrul Kilic, PhD, Gunnar P. H. Dietz, PhD, Dirk M. Hermann, MD, and Mathias Bähr, MD

623 Mitochondrial Respiratory Chain Function in Skeletal Muscle of ALS Patients

Andoni Echaniz-Laguna, MD, Joffrey Zoll, PhD, Florence Ribera, PhD, Christine Tranchant, MD, PhD, Jean-Marie Warter, MD, PhD, Jean Lonsdorfer, MD, PhD, and Eliane Lampert, MD, PhD

628 Dyskinesia after Fetal Cell Transplantation for Parkinsonism: A PET Study

Yilong Ma, PhD, Andrew Feigin, MD, Vijay Dhawan, PhD, Masafumi Fukuda, MD, Qiuhe Shi, PhD, Paul Greene, MD, Robert Breeze, MD, Stanley Fahn, MD, Curt Freed, MD, and David Eidelberg, MD

635 Neuronal and Glial Metabolite Content of the Epileptogenic Human Hippocampus

Ognen A. C. Petroff, MD, Laura D. Errante, PhD, Douglas L. Rothman, PhD, Jung H. Kim, MD, and Dennis D. Spencer, MD

Brief Communications

643 Heterozygous GLDC and GCSH Gene Mutations in Transient Neonatal Hyperglycinemia

Shigeo Kure, MD, Kanako Kojima, MD, Akiko Ichinohe, MD, Tomoki Maeda, MD, Rozalia Kalmanchev, MD, György Fekete, MD, Suzan Z. Berg, MS, Jim Filiano, MD, Yoko Aoki, MD, Yoichi Suzuki, MD, Tatsuro Izumi, MD, and Yoichi Matsubara, MD

647 Vitamin A in the Cerebrospinal Fluid of Patients with and without Idiopathic Intracranial Hypertension

Judith E. A. Warner, MD, Paul S. Bernstein, MD, PhD, Alexander Yemelyanov, MD, PhD, Stephen C. Alder, PhD, Steven T. Farnsworth, MD, and Kathleen B. Digre, MD

650 Thalamic Neurodegeneration in Multiple Sclerosis

Alberto Cifelli, MD, Marzena Arridge, PhD, Peter Jezard, PhD, Margaret M. Esiri, FRCP, Jacqueline Palace, MD, and Paul M. Matthews, FRCP

654 A Nonsense Mutation of the MASS1 Gene in a Family with Febrile and Afebrile Seizures

Junko Nakayama, MD, PhD, Ying-Hui Fu, PhD, Anna M. Clark, PhD, Satoko Nakahara, MD, PhD, Kenzo Hamano, MD, PhD, Nobuaki Iwasaki, MD, PhD, Akira Matsui, MD, PhD, Tadao Arinami, MD, PhD, and Louis J. Ptáček, MD

658 Saccadic Lateropulsion and Upbeat Nystagmus: Disorders of Caudal Medulla

Caroline Tilikete, MD, Marc Hermier, MD, Denis Pelisson, PhD, and Alain Vighetto, MD

662 Herpes Vector-Mediated Expression of Proenkephalin Reduces Bone Cancer Pain

James R. Goss, PhD, Cara F. Harley, BS, Marina Mata, MD, Mark E. O'Malley, BS, William F. Goins, PhD, Xiaoping Hu, Joseph C. Glorioso, PhD, and David J. Fink, MD

666 A New Locus for Spinocerebellar Ataxia (SCA21) Maps to Chromosome 7p21.3-p15.1

Isabelle Vuillaume, MD, David Devos, MD, Susanna Schraen-Maschke, PharmD, PhD, Christian Dina, PhD, Arnaud Lemaingue, PhD, Francis Vasseur, PhD, Guy Bocquillon, BS, Patrick Devos, BS, Carole Kocinski, BS, Christiane Marzys, BS, Alain Destée, MD, PhD, and Bernard Sablonnière, MD, PhD

670 Oligodendrocytic Polyglutamine Pathology in Dentatorubral-Pallidoluysian Atrophy

Mitsunori Yamada, MD, PhD, Toshiya Sato, MD, Shoji Tsuji, MD, PhD, and Hitoshi Takahashi, MD, PhD

675 ε-Sarcoglycan Mutations Found in Combination with Other Dystonia Gene Mutations

Christine Klein, MD, Liu Liu, BS, Dana Doheny, MS, Norman Kock, MD, Birgit Müller, MD, Patricia de Carvalho Aguiar, MD, Joanne Leung, BS, Deborah de Leon, MS, Susan B. Bressman, MD, Jeremy Silverman, PhD, Christopher Smith, MS, Fabio Danisi, MD, Chris Morrison, PhD, Ruth H. Walker, MB, ChB, PhD, Miodrag Velickovic, MD, Eberhard Schwinger, MD, Patricia L. Kramer, PhD, Xandra O. Breakefield, PhD, Mitchell F. Brin, MD, and Laurie J. Ozelius, PhD

680 "True" Sporadic ALS Associated with a Novel SOD-1 Mutation

Michael D. Alexander, MRCPI, Bryan J. Traynor, MD, Nicole Miller, PhD, Bernie Corr, RGN, Eithne Frost, MA, Shirley McQuaid, PhD, Francesca M. Brett, MD, FRCP, Andrew Green, PhD, FRCP, and Orla Hardiman, MD, FRCP

683 Symptoms in Carriers of Adrenoleukodystrophy Relate to Skewed X Inactivation

Esther M. Maier, MD, Stefan Kammerer, PhD, Ania C. Muntau, MD, Maria Wichers, MD, Andreas Braun, MD, PhD, and Adelbert A. Roscher, MD

Letters

- 689 Effect of Minocycline in Experimental Autoimmune Encephalomyelitis**
Stefan Nessler, MD, Richard Dodel, MD, Alwina Bitner, PhD, Stefanie Reuss, MD, Yansheng Du, PhD, Bernhard Hemmer, MD, and Norbert Sommer, MD
- 690 Reply**
Ian D. Duncan, BVMS, PhD, C. Linington, PhD, N. Popovic, PhD, and B. D. Goetz, BS

- 690 No Evidence for an Association between Saitohin Q7R Polymorphism and Alzheimer's Disease**
Lynnette Cook, PhD, Carol E. Brayne, MD, Douglas Easton, PhD, John Grimley Evans, MD, John Xuereb, MD, Nigel J. Cairns, MD, PhD, and David C. Rubinsztein, MB, ChB, PhD
- 691 c954C→T Polymorphism in the Fe65L2 Gene Is Associated with Early-Onset Alzheimer's Disease**
Hiroshi Tanahashi, PhD, Takashi Asada, MD, and Takeshi Tabira, MD

Regular Departments

NO 6 DECEMBER 2002

Editorials

- 695 Reversal of Diffusion Abnormalities after Ischemic Stroke: Adding Difficulty and Complexity to the Conundrum of Acute Stroke Imaging**
Marc Fisher, MD
- 696 Epilepsy and Reproductive Function in Women**
Kimford J. Meador, MD

Original Articles

- 698 Late Secondary Ischemic Injury in Patients Receiving Intraarterial Thrombolysis**
Chelsea S. Kidwell, MD, Jeffrey L. Saver, MD, Sidney Starkman, MD, Gary Duckwiler, MD, Reza Jahan, Paul Vespa, MD, J. Pablo Villablanca, MD, David S. Liebeskind, MD, Y. Pierre Gobin, MD, Fernando Vinuela, MD, and Jeffrey R. Alger, PhD
- 704 Predictors of Ovulatory Failure in Women with Epilepsy**
Martha J. Morrell, MD, Linda Giudice, MD, Kerry L. Flynn, MA, Cairn G. Seale, MS, Amelia J. Paulson, BS, Silvia Doñe, BA, Edith Flaster, MS, Michel Ferin, MD, and Mark V. Sauer, MD
- 712 Gravity Dependence of Ocular Drift in Patients with Cerebellar Downbeat Nystagmus**
Sarah Marti, MD, Antonella Palla, MD, and Dominik Straumann, MD
- 722 Accumulation of 14-3-3 Proteins in Glial Cytoplasmic Inclusions in Multiple System Atrophy**
Yasuhiro Kawamoto, MD, PhD, Ichiro Akiguchi, MD, PhD, Shinichi Nakamura, MD, PhD, and Herbert Budka, MD, PhD
- 732 Brain Alkaline Intracellular pH after Neonatal Encephalopathy**
Nicola J. Robertson, MRCP, Frances M. Cowan, PhD, I. Jane Cox, PhD, and A. David Edwards, FRCP
- 743 Frontal Lobe Atrophy Due to a Mutation in the Cholesterol Binding Protein HE1/NPC2**
Hans H. Klünemann, MD, Milan Elleder, MD, PhD, Wolfgang E. Kaminski, MD, Karen Snow, PhD, Janis M. Peyser, PhD, John F. O'Brien, MD, PhD, David Munoz, MD, Gerd Schmitz, MD, Helmfried E. Klein, MD, and William W. Pendlebury, MD
- 750 Coenzyme Q- Responsive Leigh's Encephalopathy in Two Sisters**
Lionel Van Maldergem, MD, Frans Trijbels, PhD, Salvatore DiMauro, MD, Pavel J. Sindelar, PhD, Olimpia Musumeci, MD, Antoon Janssen, MD, Xavier Delberghe, MD, Jean-Jacques Martin, MD, PhD, and Yves Gillerot, MD, PhD
- 755 Rapid Modulation of GABA in Sensorimotor Cortex Induced by Acute Deafferentation**
Lucien M. Levy, MD, PhD, Ulf Ziemann, MD, Robert Chen, MD, and Leonardo G. Cohen, MD
- 762 Selective Neurodegeneration in Murine Mucopolysaccharidosis VII Is Progressive and Reversible**
Gregory G. Heuer, PhD, Marco A. Passini, PhD, Kanli Jiang, PhD, Michael K. Parente, MS, Virginia M.-Y. Lee, PhD, John Q. Trojanowski, MD, PhD, and John H. Wolfe, VMD, PhD
- 771 Cyclooxygenase 2 Inhibition Protects Motor Neurons and Prolongs Survival in a Transgenic Mouse Model of ALS**
Daniel B. Drachman, MD, Krystl Frank, BS, Margaret Dykes-Hoberg, BS, Peter Teismann, PhD, Gabrielle Almer, MD, Serge Przedborski, MD, PhD, and Jeffrey D. Rothstein, MD, PhD
- 779 Identification of GFAP Gene Mutation in Hereditary Adult-Onset Alexander's Disease**
Michito Namekawa, MD, Yoshihisa Takiyama, MD, PhD, Yoko Aoki, MD, PhD, Norio Takayashiki, MD, Kumi Sakoe, PhD, Haruo Shimazaki, MD, Tomohiro Taguchi, MD, Yasufumi Tanaka, MD, PhD, Masatoyo Nishizawa, MD, PhD, Ken Saito, MD, PhD, Yoichi Matsubara, MD and Imaharu Nakano, MD, PhD
- 786 Changes of Sodium Channel Expression in Experimental Painful Diabetic Neuropathy**
Matthew J. Craner, MB, ChB, MRCP, Joshua P. Klein, BA, Muthukrishnan Renganathan, PhD, Joel A. Black, PhD, and Stephen G. Waxman, MD, PhD
- 793 Diet and Parkinson's Disease: A Potential Role of Dairy Products in Men**
Honglei Chen, MD, PhD, Shumin M. Zhang, MD, ScD, Miguel A. Hernán, MD, DrPH, Walter C. Willett, MD, DrPH, and Alberto Ascherio, MD, DrPH

802 Rat Forebrain Neurogenesis and Striatal Neuron Replacement after Focal Stroke

Jack M. Parent, MD, Zinaida S. Vexler, PhD,
Chao Gong, MD, PhD, Nikita Derugin, MA,
and Donna M. Ferriero, MD

814 Analysis of Pathways Mediating Preserved Vision after Striate Cortex Lesions

Mircea Ariel Schoenfeld, MD, Toemme Noesselt, PhD,
Dorothe Poggel, PhD, Claus Tempelmann, PhD,
Jens-Max Hopf, MD, Martin G. Woldorff, PhD,
Hans-Jochen Heinze, MD, and Steven A. Hillyard, PhD

Brief Communications

825 Focal Laminar Cortical MR Signal Abnormalities after Subarachnoid Hemorrhage

Jens P. Dreier, MD, Oliver W. Sakowitz, MD,
Anja Harder, MD, Claus Zimmer, MD,
Ulrich Dirnagl, MD, José M. Valdesua, MD,
and Andreas W. Unterberg, MD

829 Is the Saitohin Gene Involved in Neurodegenerative Diseases?

Patrice Verpillat, MD, Sylvain Ricard, BS,
Didier Hannequin, MD, Bruno Dubois, MD,
Jacqueline Bou, BS, Agnès Camuzat, BS,
Laurent Pradier, PhD, Thierry Frebourg, MD, PhD,
Alexis Brice, MD, PhD, Françoise Clerget-Darpoux, PhD,
Jean-François Deleuze, PhD,
Dominique Campion, MD, PhD, and the French Study
Group on Alzheimer's Disease and Frontotemporal Dementia

832 Loss of Myostatin Attenuates Severity of Muscular Dystrophy in *mdx* Mice

Kathryn R. Wagner, MD, PhD,
Alexandra C. McPherron, PhD, Nicole Winik, BS,
and Se-jin Lee, MD, PhD

836 Congenital Hypomyelinating Neuropathy, Central Dysmyelination, and Waardenburg-Hirschsprung Disease: Phenotypes Linked by *SOX10* Mutation

Ken Inoue, MD, PhD, Konstantin Shilo, MD,
Cornelius F. Boerkoel, MD, PhD, Carol Crowe, MD,
Joram Sawady, MD, James R. Lupski, MD, PhD,
and Dimitri P. Agamanolis, MD

842 Association of Chromosome 10 Losses and Negative Prognosis in Oligoastrocytomas

Lorena Bisola, PhD, Marica Eoli, MD, Bianca Pollo, MD,
Bianca Maria Merciai, PhD, Antonio Silvani, MD,
Ettore Salsano, MS, Carmelo Maccagnano, MD,
Maria Grazia Bruzzone, MD,
Anna Maria Fuhrman Conti, PhD,
Carlo Lazzaro Solero, MD, Sergio Giombini, MD,
Giovanni Broggi, MD, Amerigo Boiardi, MD,
and Gaetano Finocchiaro, MD

845 N-Acetylglutamate Synthase Deficiency and the Treatment of Hyperammonemic Encephalopathy

Orly Elpeleg, MD, Avraham Shaag, MSc,
Efrat Ben-Shalom, MD, Tal Schmid, MD,
and Claude Bachmann, MD

849 Clinical and Subclinical Dopaminergic Dysfunction in PARK6-Linked Parkinsonism: An ¹⁸F-dopa PET Study

Naheed L. Khan, MD, MRCP, Enza Maria Valente, MD,
Anna Rita Bentivoglio, MD, PhD,
Nicholas W. Wood, FRCP, PhD, Alberto Albanese, MD,
David J. Brooks, MD, DSc, FRCP, FMedSci,
and Paola Piccini, MD

853 Primary Dystonia: Is Abnormal Functional Brain Architecture Linked to Genotype?

Maja Trošt, MD, Maren Carbon, MD,
Christine Edwards, MA, Yilong Ma, PhD,
Deborah Raymond, MS, Marc J. Mentis, MD,
James R. Moeller, PhD, Susan B. Bressman, MD,
and David Eidelberg, MD

Letters

857 The Relationship of SMN to Amyotrophic Lateral Sclerosis

Thomas O. Crawford, MD,
and Richard L. Skolasky, Jr., MA

858 Reply

Philippe Corcia, MD, Bruno Moulard, MD, PhD,
Christian Andres, MD, PhD, William Camu, MD, PhD,
and the French ALS Research Group

860 Reply

Jan H. Veldink, MD,
and Leonard H. van den Berg, MD, PhD

861 Seizure-Associated Hippocampal Volume Loss: A Longitudinal Magnetic Resonance Study of Temporal Lobe Epilepsy

Rebecca S. N. Liu, BSc, MRCP, Louis Lemieux, PhD,
Josemir W. A. S. Sander, PhD, MRCP,
Sanjay M. Sisodiya, PhD, MRCP,
and John S. Duncan, DM, FRCP

862 Reply

Regula S. Briellmann, MD, Ari Syngeniotis,
Samuel F. Berkovic, and Graeme D. Jackson

Regular Departments

863 Books

865 American Neurological Association, Works in Progress Abstracts

867 American Neurological Association, Summer Student Research Abstracts

869 American Neurological Association, Call for Abstracts, Neurology Outcomes Research

Indexes to Volume 52

870 Author Index

882 Subject Index